A native mitral valve mass beyond imagination

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Abstract

The authors report a case of a patient with a history of immunoglobulin A nephropathy who, during the admission for pneumonia, had an incidental finding of a huge mitral valve (MV) mass on transthoracic echocardiography. The differential diagnosis was challenging because the clinical scenario raised the suspicion of possible infective endocarditis, and the imaging features were suggestive of a myxoma or vegetation. The patient underwent urgent excision of the mass with MV replacement due to the high risk of embolism. Intraoperative findings were consistent with clots or vegetation. The pathology result of the thrombus was beyond our imagination, and to the best of our knowledge, only one case has been reported. Awareness about native MV thrombosis and its etiologic factors, work-up, and management is key for better medical and surgical management planning because this condition is extremely rare and challenging in the clinical and imaging arenas.

Case Report

A year before admission, a 32-year-old male with a history of hypertension experienced shortness of breath (SOB), hemoptysis, and lower limb edema after receiving the COVID-19 vaccine. He was hospitalized and diagnosed with acute kidney injury due to immunoglobulin A (IgA) nephropathy, confirmed by a renal biopsy. The patient did not have a history of tuberculosis or intravenous drug use but had a family history of systemic lupus erythematosus.

One month before his admission to our hospital, the patient sought treatment at a local hospital for new-onset SOB, chest pain, and hemoptysis lasting 2 weeks. He was diagnosed with pneumonia and treated with antibiotics. During routine transthoracic echocardiography (TTE), a mass on the mitral valve (MV) was discovered, prompting a referral to our hospital. The patient's work-up showed an electrocardiogram of normal sinus rhythm, elevated creatinine levels (321 umol/L), normal complete blood counts, and negative blood cultures. Immunological tests revealed an increased erythrocyte sedimentation rate as well as positive anti-cardiolipin antibodies. Coagulation profile tests indicate a D-dimer measurement of 0.58 ug/mL, fibrinogen levels of 8.2 g/L (elevated), a partial thromboplastin time (PTT) reading of 89 seconds (elevated), and a normal international normalized ratio. Testing for Goodpasture syndrome was negative. TTE showed a multilobulated, large, raceme-shaped, mobile mass protruding in the diastole into the left ventricle (LV) with irregular borders. It was a non-homogeneous, mainly myocardial-like texture with focal areas of hyperechogenicity (size 19×19 mm), likely attached by a broad peduncle on the atrial side of the basal mid portion of the anterior mitral leaflet (AML) (Figure 1).

The MV leaflets were thickened with focal areas of calcification. There was an eccentric systolic jet directed toward the posterolateral atrial wall due to the AML-impaired mobility (mass-related), causing incomplete coaptation of the leaflets and severe mitral regurgitation. The LV was normal in size and function. A contrast TTE study using Optison infusion was performed to detect the blood supply of the mass, and it showed no early or 20-minute delayed uptake into the mass (Figure 1).

Further 2D and 3D transesophageal echocardiography (TEE) allowed a better visualization and sizing of the mass (found larger than in TTE, size 3.2×1.1 cm). TEE confirmed the sessile attachment to the base/mid portion of the atrial surface of the AML. 2D and 3D TEE allowed us to better delineate the morphologic features of the mass (site and type of attachment, texture, shape) (Figure 2).

The case was discussed during the multi-disciplinary team (MDT) meeting. According to the clinical and imaging patterns, it was decided to proceed with emergent surgery due to the high risk of embolism even without further imaging work-up by magnetic resonance imaging (MRI). The patient was maintained on a heparin infusion until the time of the surgery. The preoperative TEE revealed no change in size.

At surgery, MV repair was not possible (due to the calcifications of the leaflets), and the mass was impossible to resect as it was strongly attached without damaging the leaflets. Therefore, the MV was replaced. We found a red, mushy tissue between the gelatinous and firm tissue, with an irregular surface. Its size was about 3.4 cm. The mass was attached to the atrial surface of the AML (Figure 3).

The surgeon's impression of the anatomical features of the lesion (consistency, color, surface) was of vegetation or clot. The patient had an uneventful surgical course and was discharged home in stable condition. After immunohistochemistry and histological analysis of the mass, it was found to be a thrombus.

Discussion

Without rheumatic heart disease or thrombophilic disease, native MV leaflet thrombosis is extremely rare, and to the best of our knowledge, only one case has been reported. Prior cases of native valve thrombi have been associated with antiphospholipid syndrome, hypercoagulable states, hypereosinophilic syndrome, and rheumatic MV [1-6]. They more often involve prosthetic valves, with an incidence of 1% to 3% per year [7-9]. We report a rare clinical case of a patient with a history of IgA nephropathy, pneumonia, and a huge mass on the native MV. The diagnosis was challenging because the clinical scenario raised the suspicion of possible infective endocarditis [7], while the patient was found to have an MV mass during concomitant pneumonia and the imaging features were suggestive of a myxoma or vegetation [10-15], the mass being huge in size, raceme-shaped, and attached to a leaflet with normal mobility, therefore an unlikely site of origin of a large clot in particular without a history of known thrombophilic disease.

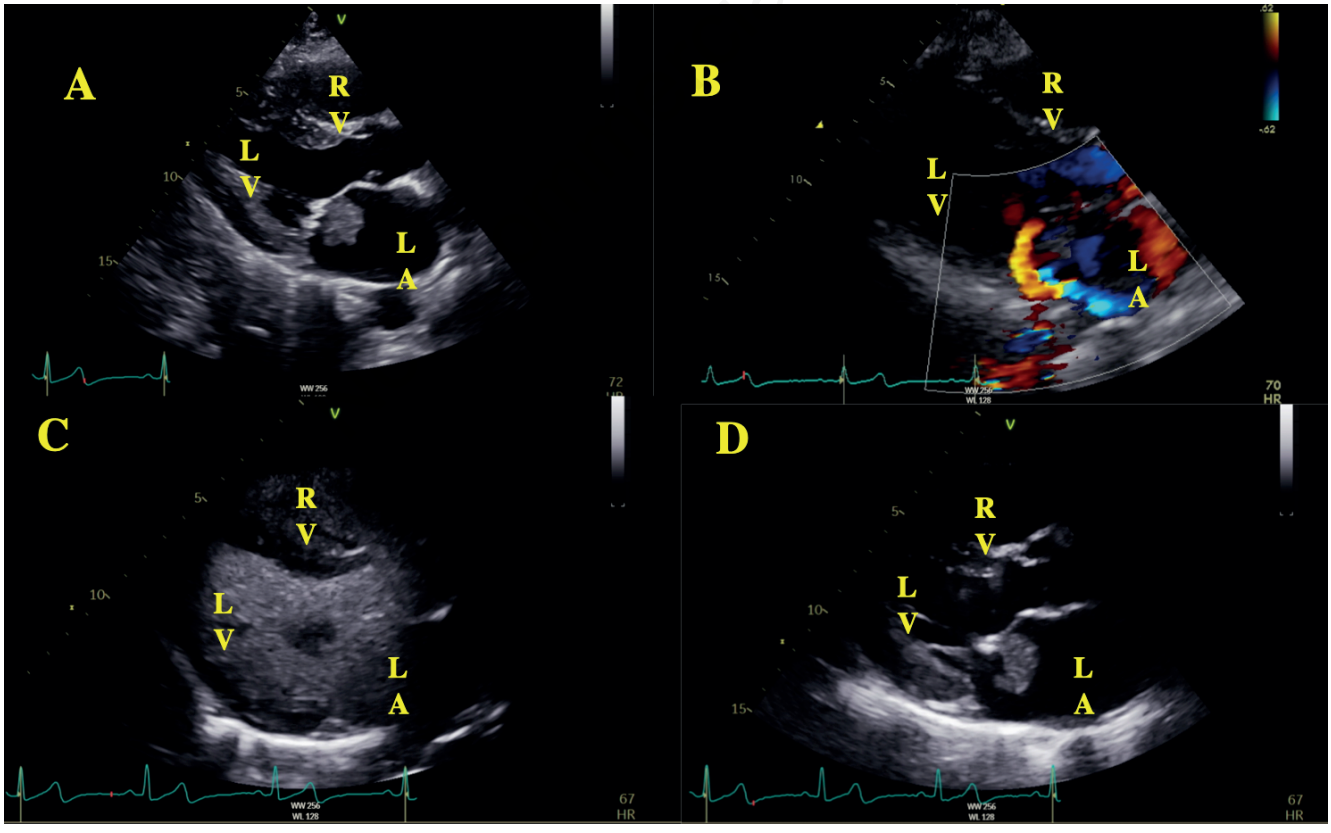

Figure 1. Trans-thoracic echocardiography. A) Parasternal long axis view (PLAX) at mid-systole showing the mass attached to the anterior mitral leaflet; B) PLAX, systole, color doppler at mitral valve showing posteriorly directed jet of mitral regurgitation; C,D) baseline contrast-enhanced echocardiography by Optison showing left ventricular opacification and no uptake of contrast the mass (baseline) (C) and 20 minutes after the contrast (D). LV, left ventricle; LA, left atrium; RV, right ventricle.

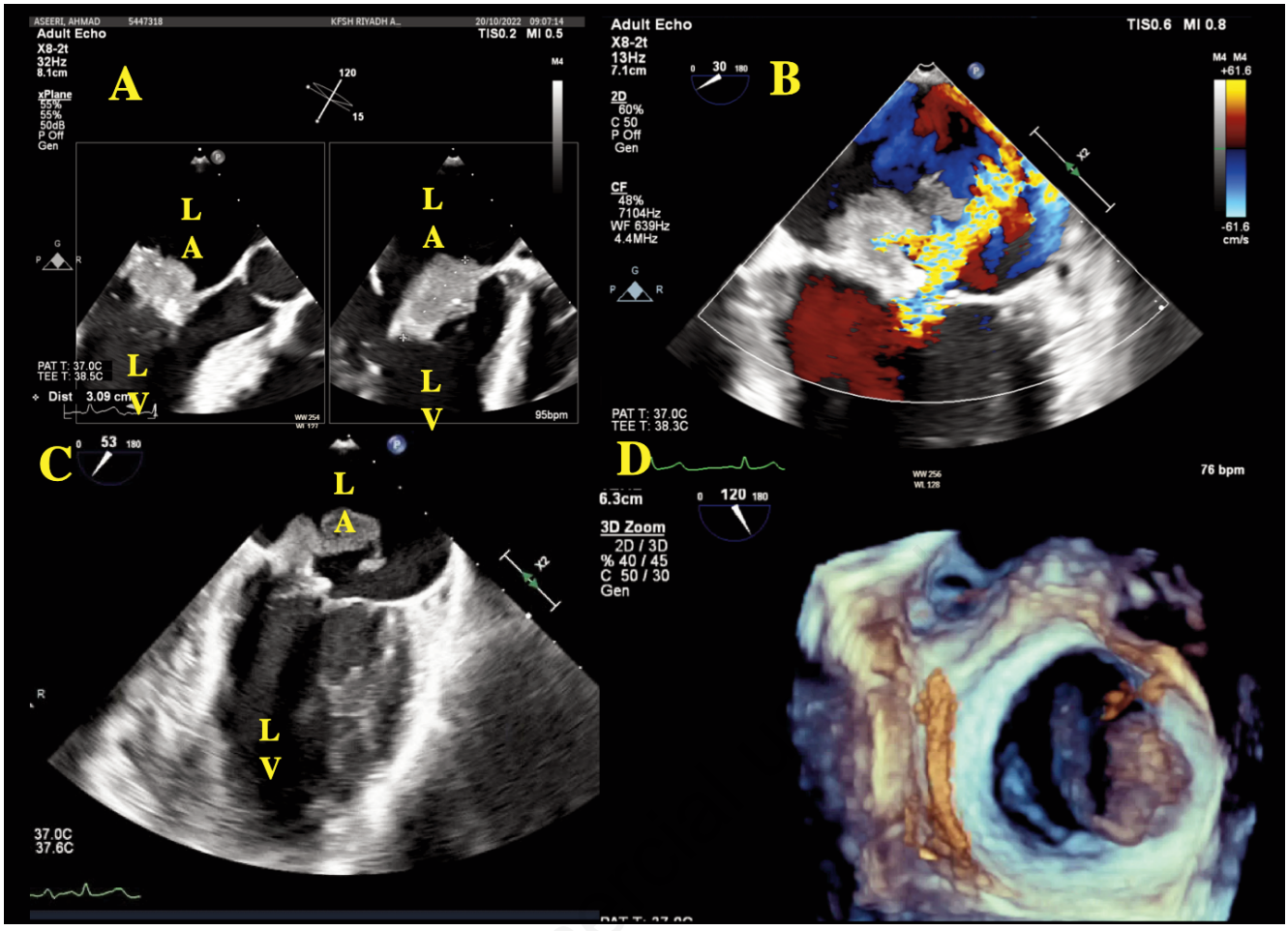

Figure 2. Transesophageal echocardiography. A) Mid-esophageal view at 120 degrees with X-plane orthogonal views in diastole showing the mass attached to the anterior mitral leaflet prolapsing into the left ventricle; B) mid-esophageal view, color Doppler across the mitral showing eccentrically directed jet and severe mitral regurgitation; C) bi-commissural view at mid-systole showing the mass attached to the A2 segment; D) 3D surgical view of the mitral valve showing the mass (attached to the atrial surface of anterior mitral leaflet). LV, left ventricle; LA, left atrium.

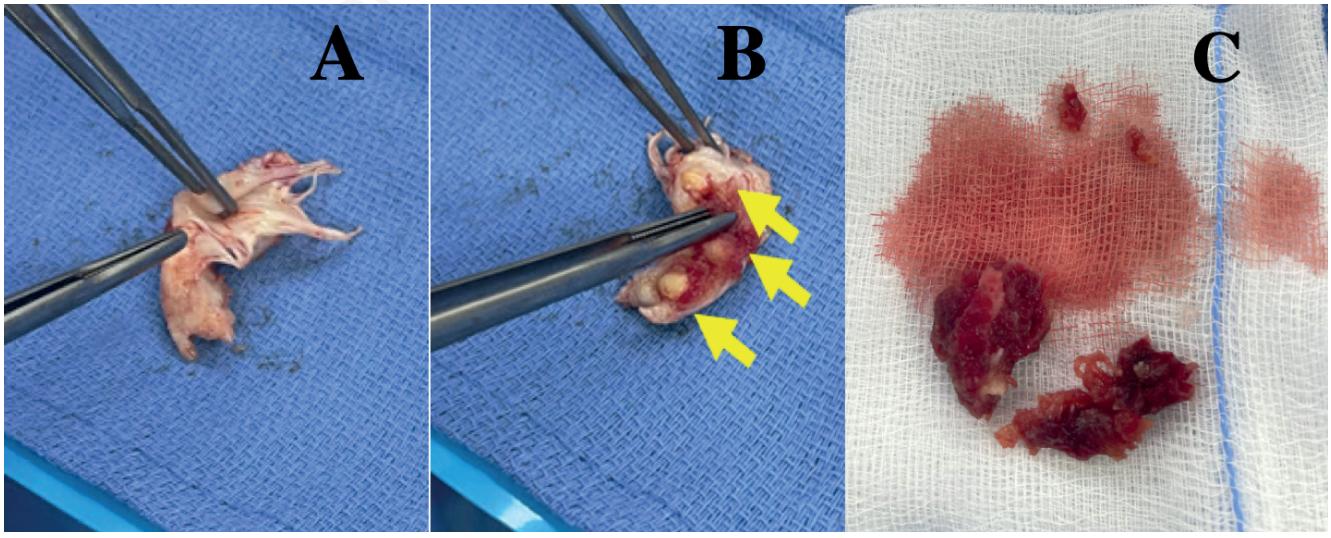

Figure 3. Surgical specimen. A) Specimen of the anterior mitral leaflet, ventricular surface; B) anterior mitral leaflet side calcified nodule (yellow arrow); C) red, mushy tissue between the gelatinous and firm tissue, irregular surface size 3.4 cm mass (thrombus).

The possibility of remnants of healed vegetation was also unlikely due to the size of the mass. Our MDT meeting consensus was that the very high risk of embolism required proceeding with emergent surgery without any further MRI imaging and laboratory work-up. However, we have to highlight that, in the intracardiac mass imaging pathway, MRI has a key role. Even though contrast echocardiography did not show any early or late uptake, therefore suggesting a structure without blood supply [10], this finding was believed to suggest vegetation more than clots. The intraoperative findings were consistent with either vegetation or clot (appearance, color, consistency, and adherence). Pathological and histochemical evaluation of the mass and the leaflets unexpectedly found the mass to be a clot with no findings of infective endocarditis or cellular transformation. Clot had not been considered because thrombosis rarely occurs on native valves. In the intracardiac mass imaging pathway, echocardiography represents the first diagnostic technique able to diagnose the presence of an intracardiac mass and visualize the anatomical features of the lesion [10-12]. In the imaging arena, 3D TEE can have an important adjunctive value, as it is better able to demonstrate many anatomical features and in particular the type and site of attachment of pedunculated cardiac tumors [10-15]. In our case, 3D TEE provided the best visualization of the attachment. Cardiac magnetic resonance has a key role in the diagnosis of cardiac mass and is now a highly effective and powerful tool due to its soft tissue characterization sequences, including fat saturation, water content, blood perfusion, vascularity of the mass, as well as anatomical features including attachment and size [13].

Prior cases of native valve thrombi have been associated with antiphospholipid syndrome, hypercoagulable states, hypereosinophilic syndrome, and rheumatic MV [1-7]. Only one case of thrombosis on a native non-rheumatic MV has been reported [1].

It has been reported that patients with chronic kidney disease (CKD) manifest a coagulopathy consisting of delayed clot formation but increased final clot strength and decreased clot breakdown [15,16]. The increased clot strength is mediated by elevated fibrinogen levels in CKD. The findings of delayed clot formation, decreased lysis, and increased fibrinogen levels have been reported in previous studies. Although the delayed clot formation seen may predispose to bleeding complications, the increased clot strength and decreased breakdown in this group may account for the increased thrombotic complications in this group. Thus, methods to treat hypercoagulability in this population should also consider targeting fibrinogen [13,16,17].

In our case, the patient was affected by IgA nephropathy; the coagulation profile showed a positive D-dimer of 0.58 ug/mL; an increased fibrinogen level of 8.2 g/L, 89-second PTT; and anti-cardiolipin antibodies were positive, supporting the presence of a hypercoagulable state. However, the pathogenetic mechanism of the formation of a huge clot on the native MV leaflet is very unclear. In fact, in our case, there was no atrio-ventricular low-flow status that is present in conditions like rheumatic valvulopathies, arrhythmias, or reduced left ventricular function that can predispose to the formation of large clots [18].

We speculated as a possible pathogenetic pathway a clot formation on a damaged, inflamed leaflet surface due to a possible valvulitis either by infective endocarditis or immunological phenomena on a degenerative leaflet because of CKD (the MV leaflet was thickened and with extensive calcification) in a patient that has a hypercoagulable state due to a post-COVID-19 vaccine IgA nephropathy.

While one case had been reported on the MV, thrombosis on the native aortic valve, which is also uncommon, is relatively more frequent. In a recent meta-analysis, 74 cases of aortic valve thrombosis were reported. The most common underlying etiologies were hypercoagulable diseases (30%), idiopathic diseases (19%), left ventricular assist devices (18%), aortic valve or root disease (17%), and congenital heart disease (8%). This condition appeared to be associated with an increased risk of poor in-hospital outcomes. Therefore, aortic valve thrombosis is more clinically relevant in patients with embolic events [19].

The scenario of a clot on native MV can also widen the horizons towards different therapeutic strategies by thrombolysis that were not considered in our clinical scenario, different modalities of anticoagulation, surgical timing, and, when surgical resection is believed necessary for the high risk of embolism, the repair of the valve as the best option. However, in our case, urgent surgery due to the very high risk of embolism in the setting of possible infective endocarditis in a young male was considered the best therapeutical choice [7], and the repair was not feasible due to the degenerative MV and the size and anatomical features of the mass.

Conclusions

Awareness about native MV thrombosis and its etiologic factors, diagnostic work-up, and management is paramount for better medical and surgical management planning because this condition is extremely rare and challenging in the clinical and imaging arena and can be associated with a poor outcome.

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